

REVISION OF PREVIOUS FONTAN CONNECTIONS TO TOTAL EXTRACARDIAC CAVOPULMONARY ANASTOMOSIS: A MULTICENTER EXPERIENCE

Carlo F. Marcelletti, MD^a
 Frank L. Hanley, MD^b
 Constantine Mavroudis, MD^c
 Doff B. McElhinney, MD^b
 Raul F. Abella, MD^a
 Stefano M. Marianeschi, MD^a
 Francesco Seddio, MD^a
 V. Mohan Reddy, MD^b
 Ed Petrossian, MD^b
 Teresa de la Torre, MD^a
 Luisa Colagrande, MD^a
 Carl L. Backer, MD^c
 Adriano Cipriani, MD^a
 Fiore S. Iorio, MD^a
 Francis Fontan, MD^d

Background: Conversion to total extracardiac cavopulmonary anastomosis is an option for managing patients with dysfunction of a prior Fontan connection. **Methods:** Thirty-one patients (19.9 ± 8.8 years) underwent revision of a previous Fontan connection to total extracardiac cavopulmonary anastomosis at four institutions. Complications of the previous Fontan connection included atrial tachyarrhythmias ($n = 20$), progressive heart failure ($n = 17$), Fontan pathway obstruction ($n = 10$), effusions ($n = 10$), pulmonary venous obstruction by an enlarged right atrium ($n = 6$), protein-losing enteropathy ($n = 3$), right atrial thrombus ($n = 2$), subaortic stenosis ($n = 1$), atrioventricular valve regurgitation ($n = 3$), and Fontan baffle leak ($n = 5$). Conversion to an extracardiac cavopulmonary connection was performed with a nonvalved conduit from the inferior vena cava to the right pulmonary artery, with additional procedures as necessary. **Results:** There have been 3 deaths. Two patients died in the perioperative period of heart failure and massive effusions. The third patient died suddenly 8 months after the operation. All surviving patients were in New York Heart Association class I ($n = 20$) or II ($n = 7$), except for 1 patient who underwent heart transplantation. Early postoperative arrhythmias occurred in 10 patients: 4 required pacemakers, and medical therapy was sufficient in 6. In 15 patients, pre-revision arrhythmias were improved. Effusions resolved in all but 1 of the patients in whom they were present before revision. The condition of 2 patients with protein-losing enteropathy improved within 30 days. **Conclusions:** Conversion of a failing Fontan connection to extracardiac cavopulmonary connection can be achieved with low morbidity and mortality. Optimally, revision should be undertaken early in symptomatic patients before irreversible ventricular failure ensues. (J Thorac Cardiovasc Surg 2000;119:340-6)

In 1971, Professor Fontan first constructed a completely in-series circulation with exclusion of the right ventricle, using the right atrium as a dynamic element, in a patient with tricuspid atresia.¹ Since this monu-

mental event, the physiologic understanding of the Fontan circulation has been evolving steadily, with various modifications and expanding applications in patients with univentricular heart disease. Patients who have undergone these or further modifications of the Fontan procedure may have progressive exercise intolerance, atrial arrhythmias, recurrent effusions, and protein-losing enteropathy, as well as other manifestations of chronic congestive failure and a malfunctioning atrio-pulmonary connection.²⁻⁴ The increase in right atrial size and pressure that occurs in the setting of an atrio-pulmonary Fontan connection has been hypothesized to cause turbulence and energy loss, compression of the pulmonary veins, coronary sinus hypertension, arrhythmias, and atrial thrombus.⁵⁻⁷

Few therapeutic options are available for patients having these problems. Management is for the most part

From the Hesperia Hospital,^a Modena, Italy; University of California,^b San Francisco, Calif; Children's Memorial Hospital,^c Chicago, Ill; and Clinique St Augustin,^d Bordeaux, France.

Read at the Seventy-ninth Annual Meeting of The American Association for Thoracic Surgery, New Orleans, La, April 18-21, 1999.

Received for publication April 22, 1999; revisions requested June 25, 1999; revisions received Aug 10, 1999; accepted for publication Oct 6, 1999.

Address for reprints: Carlo F. Marcelletti, MD, Pediatric Cardiac Surgery, Hesperia Hospital, Via Arquà 80/A, Modena, 41100 Italy (E-mail: marianeschi@hesperia.it).

Copyright © 2000 by Mosby, Inc.

0022-5223/2000 \$12.00 + 0 12/6/103465

limited to orthotopic heart transplantation or revision of the Fontan connection. Revision to a lateral tunnel has been described by several investigators,⁷⁻⁹ and in recent years a number of centers have adopted conversion to an extracardiac conduit total cavopulmonary anastomosis as the therapy of choice for patients with Fontan failure.¹⁰⁻¹² The aim of the current study is to present a multicenter experience with conversion of previous Fontan connections to total extracardiac cavopulmonary anastomosis, including patients undergoing revision at the Hesperia Hospital in Modena (Italy), the University of California at San Francisco (California), the Children's Memorial Hospital in Chicago (Illinois), and the Clinique St Augustin in Bordeaux (France).

Patients and methods

Patients. Between October 1992 and January 1999, 31 patients between 6 and 36 years of age (mean \pm SD 19.9 \pm 8.8 years) underwent conversion of an atriopulmonary (n = 29), atrioventricular (n = 1), or lateral tunnel (n = 1) Fontan connection to total extracardiac cavopulmonary anastomosis at the four institutions listed above. Some of these patients have been included in previous reports.¹⁰⁻¹² The interval between the original Fontan procedure and conversion to an extracardiac cavopulmonary anastomosis ranged from 6 to 20.5 years (mean \pm SD 12.1 \pm 4.8 years). Three patients had undergone a revision of the Fontan procedure (replacement of the valved conduit with a nonvalved synthetic tube) from 8.1 to 12 years after the original procedure and from 6 to 10 years before the conversion. All patients had undergone palliative shunting procedures or classic superior cavopulmonary anastomosis before the initial Fontan operation. Primary diagnoses were univentricular transposition complexes (n = 10), tricuspid atresia with or without pulmonary stenosis/atresia (n = 11), double-inlet left ventricle (n = 5), double-outlet right ventricle and mitral atresia (n = 2), pulmonary atresia and intact ventricular septum (n = 2), and unbalanced complete atrioventricular septal defect (n = 1). Echocardiography and cardiac catheterization were performed before revision in all patients.

Patients had a wide range of complications of the original Fontan connection. All patients were clinically symptomatic, with 13 patients in New York Heart Association (NYHA) class IV, 12 in class III, and 6 in class II. Ventricular function measured by echocardiography was considered good or only mildly decreased in 20 patients and poor in 11 patients. At preoperative catheterization, the ventricular end-diastolic pressure ranged from 2 to 13 mm Hg (mean \pm SD 7.4 \pm 3.6 mm Hg). Atrial arrhythmias were present in almost all patients, although these were not hemodynamically or clinically significant in all cases: 10 patients had supraventricular tachycardia causing hemodynamic compromise or syncopal episodes, necessitating repeated hospitalizations and multiple antiarrhythmic medications; 9 patients had atrial flutter; 1 had atrial fibrillation; 3 had first-degree atrioventricular block;

and 5 had previously undergone implantation of a permanent bipolar epicardial pacemaker because of atrioventricular block (n = 4) or sick sinus syndrome (n = 1). Three patients did not have documented rhythm disturbances. Pleural and pericardial effusions were present in 10 patients, and 3 patients had ascites and protein-losing enteropathy necessitating long-term albumin supplementation. The cardiac index was less than 2 L \cdot min⁻¹ \cdot m⁻² in 10 patients. Compression of the pulmonary veins by the enlarged right atrium was documented by 2-dimensional echocardiography in 6 patients. The mean right atrial pressure was 15.2 mm Hg and was 20 mm Hg or more in 5 patients, 2 of whom had a large thrombus filling the right atrium. Pulmonary artery pressure averaged 13.2 \pm 5.2 mm Hg. Obstruction of the Fontan pathway (pressure gradient \geq 3 mm Hg between the superior vena cava and pulmonary artery) was present in 14 patients. Significant atrioventricular valve regurgitation was present in 8 patients, with moderate or severe regurgitation in 3 and mild regurgitation in 5. The mean \pm SD systemic oxyhemoglobin saturation was 88% \pm 5%. Nine patients had arterial oxygen saturations less than 90%, 5 of whom had a baffle leak or partial patch detachment. None had fenestrations before revision. Four patients had ventilation/perfusion ratio mismatch because of pulmonary arteriovenous malformations.

Operative techniques. Careful redo median sternotomy was the surgical approach. Standard techniques of cardiopulmonary bypass were used, with aortic and bicaval cannulation and moderate hypothermia (28°C-32°C). Seven patients were placed on cardiopulmonary bypass by means of femoro-femoral cannulation and then transitioned to aortic and bicaval cannulation after successful sternotomy and exposure of the heart and great vessels. After institution of bypass, the aorta was crossclamped and cold crystalloid or blood cardioplegic solution was given into the aortic root. The technique of cardiopulmonary bypass was modified as necessary according to the additional anomalies. In 6 patients, deep hypothermic circulatory arrest was used.

Conversion to a total extracardiac cavopulmonary connection was carried out with a conduit from the inferior vena cava to the right pulmonary artery. A nonvalved polytetrafluoroethylene tube graft was used in 25 patients, an allograft conduit in 3, and a Dacron tube in 3. The diameter of the conduit was matched to that of the inferior vena cava and ranged from 20 to 25 mm. In patients without a previously placed bidirectional superior cavopulmonary connection, either this procedure was performed or a prior classic Glenn shunt was incorporated into the extracardiac Fontan circuit. In 1 patient, an aortic allograft was used to restore pulmonary arterial continuity. In 21 patients (68%), reduction right atrioplasty was performed on the anterior right atrial wall, away from the conduction tissue; the crista terminalis was not resected. Reconstruction or augmentation (or both) of the pulmonary arteries was carried out in 11 patients. In 6 patients, intraoperative mapping and cryoablation of arrhythmia circuits was performed and an antitachycardia pacemaker was placed. In 2 other patients, both of whom underwent reduction right

atrioplasty, a modified maze procedure was performed. Two patients required right atrial thrombectomy and 2 underwent ligation of a left superior vena cava. In 1 patient, a severely regurgitant tricuspid valve was closed with a patch. In the other patients with atrioventricular valve regurgitation before revision, no procedures were performed on the valve. In 1 patient, the bulboventricular foramen was enlarged through the right atrium and aortic valve by resecting a wedge of tissue from the apical trabecular septum, which was incised close to the obtuse margin of the ventricular mass. No other intracardiac procedures were performed and no atrial/conduit fenestrations were used.

Results

Operative and early postoperative results. There were 2 operative deaths. A 25 year-old patient died of irreversible congestive heart failure shortly after the operation. Preoperatively, the patient had effusions, compression of the right pulmonary veins, moderate to severe mitral regurgitation, and was in NYHA functional class IV. This patient tolerated the procedure well and was in stable condition with the support of inotropic drugs. However, after sternal closure the patient had a cardiac arrest and did not respond to resuscitative efforts. The second perioperative death occurred in a patient with severe cachexia and multiple complications of the Fontan circulation, in whom transplantation was contraindicated for social reasons. This patient (who had severe effusions, protein-losing enteropathy, and congestive heart failure before the operation) experienced massive effusions with increasing acidosis. On the fourth postoperative day, he was returned to the operating room, where in a salvage attempt the Fontan connection was taken down to a bidirectional cavopulmonary anastomosis with a supplemental modified Blalock-Taussig shunt. The hemodynamic status continued to decline, and shortly after reoperation the patient had a cardiac arrest and could not be resuscitated.

Five other patients underwent early reoperation: 1 patient required reoperation for cardiac tamponade due to chronic pericardial effusion, and 4 had permanent dual-chamber epicardial pacemakers placed. The remaining patients required postoperative mechanical ventilation for a median of 13 hours (7-29 hours) and inotropic medications for a median of 3 days (1-5 days). Effusions resolved within 30 days in all surviving patients in whom they were present before revision, but 1 patient had significant new effusions develop, necessitating chest tube drainage for 21 days. The condition of 2 patients with protein-losing enteropathy improved, and within 30 days their serum albumin lev-

els increased without exogenous replacement to 3.5 and 4.5 g/dL from 2 and 2.3 g/dL, respectively. In addition to the patients who had placement of a permanent epicardial pacemaker, 6 patients had persistence of pre-revision arrhythmias, which were controlled effectively with medical therapy. One patient who did not have preoperative rhythm disturbances had atrial flutter/fibrillation develop after the operation. In 15 patients, pre-revision arrhythmias were improved. In 9 of these patients, the arrhythmias disappeared, and in 6 they became responsive to medical therapy whereas they were unresponsive before the operation. All but 1 of these patients had undergone reduction right atrioplasty. Arrhythmias were controlled in all of the patients who underwent intraoperative cryoablation except for 1, who had recurrent supraventricular tachycardia that was controlled with procainamide. Of the 29 early survivors, 24 were discharged receiving warfarin sodium therapy (international normalized ratio maintained in the 1.5 to 2.0 range), and the remaining 5 were placed on an aspirin regimen.

Follow-up. Cross-sectional follow-up was obtained in all hospital survivors, with a median duration of 25 months (4-61 months). One death occurred during the follow-up period in a patient who died 8 months after the operation of a malignant arrhythmia. Preoperatively, this patient had a permanent pacemaker because of atrioventricular block, and in the early postoperative period this patient did not manifest any new arrhythmia activity. No autopsy was performed. One patient had recurrent, refractory atrial flutter and progressive ventricular dysfunction, necessitating placement of a pacemaker 8 months after the operation and orthotopic heart transplantation 18 months after revision. At follow-up 10 months later, the patient is alive and in stable condition in NYHA class I. One patient required late reoperation for wound infection 2 months after revision.

At most recent follow-up, all surviving patients were in NYHA functional class I (n = 20) or II (n = 7) except for the patient who required heart transplantation. This patient was initially in NYHA class II after revision, deteriorated to class IV, and after transplantation was in NYHA class I. All patients had follow-up echocardiography, which demonstrated improvement in atrioventricular valve regurgitation in both patients with moderate to severe regurgitation who did not undergo procedures on the atrioventricular valve at the time of reintervention. New atrioventricular or semilunar valve regurgitation had not developed in any patient. No patients had evidence of a decline in ventricular function except for the patient who underwent heart trans-

plantation. In the remaining patients, ventricular function was normal or unchanged after revision, with significantly improved systolic and diastolic function in 4 patients. Follow-up catheterization was performed in 6 patients.

Discussion

Few therapeutic options are available for patients with a Fontan connection and deteriorating functional status. Conservative medical therapy may be sufficient if the patient is in reasonably functional condition. Several investigators have reported orthotopic heart transplantation as management options for patients with end-stage disease.⁸⁻¹² However, there is a group of patients with deteriorating functional status in whom a better first option may be conversion of the failing Fontan circuit to total cavopulmonary connection. Specific indications for an optimal timing of this approach are not clearly defined. The most obvious candidates for conversion are patients with complications that stand to be addressed directly by revision, including a giant atrium with thrombus, arrhythmias refractory to medical or simple electrophysiologic treatment, pulmonary venous obstruction by the enlarged right atrium, and ventricular dysfunction resulting from elevated coronary sinus pressure.^{9,11,12} Patients with intrinsic myocardial dysfunction are less likely to benefit from conversion, although select patients in this category may benefit. It can be difficult to determine the extent to which the ventricular dysfunction is related to arrhythmias and/or coronary sinus hypertension, which may be in part reversible.

Among the several modifications of the Fontan procedure to which a failing connection may be converted, we believe that total cavopulmonary anastomosis with an extracardiac conduit is a good option in most instances. Although several investigators have reported improvement after conversion to an intracardiac lateral tunnel type connection,⁷⁻⁹ an extracardiac total cavopulmonary anastomosis may be preferable in the long run because of several potential advantages. Not only is an extracardiac Fontan operation easier and more flexible to perform, especially in patients with extensive prior surgery, but it can be expected to result in superior hydrodynamic efficiency¹³ and allows more aggressive strategies to minimize residual or recurrent complications related to the right atrium. The extracardiac conduit approach avoids additional atrial suture lines (except for that required to close the reduction atrioplasty) and allows more extensive reduction right atrioplasty, which not only eliminates the dysfunction and arrhythmogenic right atrial tissue in the Fontan

pathway, but provides a convenient bed for placement of the conduit. There are numerous other potential benefits of extracardiac total cavopulmonary anastomosis, which have been detailed elsewhere and will not be elaborated on in this discussion.^{14,15}

In this multicenter experience with 31 patients, the two principal indications for conversion were arrhythmias and hydrodynamic inefficiency of the Fontan circuit, with atrial tachyarrhythmias in 20 patients and progressive congestive heart failure in 17. Although patients with specific complications of the previous Fontan procedure are most likely to benefit from revision, the utility of conversion in patients with deteriorating functional status and no specific target conditions is questionable. This would include patients with significantly depressed ventricular function, as well as patients with very limited functional status who have moderately elevated pulmonary vascular resistance, moderately depressed ventricular function, a large right atrium without specific complications, and no obstructive lesion. This group of highly debilitated patients may not tolerate the insult of the operation itself. However, they may not tolerate transplantation either, and the relative risks and benefits of the two approaches should be assessed. It should be kept in mind, however, that ventricular function may be improved in patients with less significant impairment simply by decreasing the pressure in the right atrium and coronary sinus, which is illustrated by the fact that many of our patients had a reduction in ventricular end-diastolic pressure after revision.

All of the patients who survived the perioperative period had an immediate clinical improvement after conversion. The only perioperative deaths were in especially ill patients, who were suboptimal candidates for revision but in whom this was the only realistic option. In general, the particular complications (such as obstruction of the Fontan pathway or pulmonary veins, arrhythmias, and effusions) were rectified, presumably restoring some degree of integrity to the Fontan circuit. In many patients, at least part of the improvement could be traced to effective control of previously intractable arrhythmias. Although 4 patients had a new pacemaker placed at the time of revision (not including the 6 in whom intraoperative cryoablation and placement of an antitachycardia pacemaker was performed), arrhythmias were improved substantially in 15 patients. Six patients were treated intraoperatively with cryoablation, 2 had a modified maze procedure, and most underwent reduction right atrioplasty, which should be considered almost an essential component of conversion of a failing Fontan connection to extracardiac con-

duit cavopulmonary anastomosis. This applies to patients without as well as with preoperative arrhythmias. Chronic right atrial hypertension and resulting distention alters the electrical characteristics of the atrial myocardium, increasing its susceptibility to arrhythmogenesis. Even if the tachyarrhythmias characteristic of a failing Fontan shunt have yet to manifest themselves in a given patient, the threshold for such arrhythmias in the perioperative period is likely to be lowered, and reduction right atriotomy is likely to be beneficial. Moreover, these patients may have some degree of obstruction of the right pulmonary veins because of compression by the dilated right atrium. This complication was diagnosed in 20% of the patients in this series and was likely present to a less obvious extent in others as well. Resection of dilated right atrial free wall may help to eliminate this and other potential mass effects and provides additional space in the mediastinum for placement of the conduit. Although none of the patients in this series had fenestration of the extracardiac cavopulmonary connection, fenestration may help reduce the duration of effusions in some patients. However, post-conversion effusions were not a common complication in the present series, and we do not have sufficient data to recommend or not recommend fenestration in this group of patients.

A frequent and vexing complication of the modified Fontan procedure is protein-losing enteropathy. The causes of this complication are not well understood, nor are the factors that may contribute to effective therapy. Two of 3 patients with this complication in the present cohort improved within 30 days, presumably because of altered hemodynamic conditions. The number of patients with protein-losing enteropathy in this series is too few to recommend revision specifically for this complication, but substantial improvement in 2 of 3 patients suggests that it may be a worthwhile option in appropriate patients. Further investigation is warranted to determine which patients with this complication may benefit from conversion to total extracardiac cavopulmonary connection.

In conclusion, revision of the failing Fontan to extracardiac conduit total cavopulmonary connection can be performed with significant improvement in most patients and low mortality (10%) and morbidity. Optimal selection criteria for conversion have yet to be determined, but most patients can be expected to benefit to some extent. The degree of improvement is likely to correspond with the proportion of complications of the previous Fontan connection in a given patient that can be addressed by conversion. The significance of these cumulative results is profound, considering that

the only alternative for many of these patients is cardiac transplantation.

REFERENCES

1. Fontan F, Baudet E. Surgical repair of tricuspid atresia. *Thorax* 1971;26:240-8.
2. Peters NS, Somerville J. Arrhythmias after the Fontan procedure. *Br Heart J* 1992;68:199-204.
3. Gelatt M, Hamilton RM, McCrindle BW, et al. Risk factors for atrial tachyarrhythmias after the Fontan operation. *J Am Coll Cardiol* 1994;24:1735-41.
4. de Leval MR, Kilner P, Gewillig M, Bull C. Total cavopulmonary connection: a logical alternative to atriopulmonary connection for complex Fontan operations. *J Thorac Cardiovasc Surg* 1988;96:682-95.
5. Miura T, Hiramatsu T, Forbess JM, Mayer JE. Effects of elevated coronary sinus pressure on coronary blood flow and left ventricular function: implications after the Fontan operation. *Circulation* 1995;92(Suppl):II298-303.
6. de Leval MR. Right heart bypass operations. *Semin Thorac Cardiovasc Surg* 1994;6:8-12.
7. Kao JM, Alejos JC, Grant PW, Williams RG, Shannon KM, Laks H. Conversion of atriopulmonary to cavopulmonary anastomosis in management of late arrhythmias and atrial thrombosis. *Ann Thorac Surg* 1994;58:1510-4.
8. Kreutzer J, Keane JF, Lock JE, et al. Conversion of modified Fontan procedure to lateral atrial tunnel cavopulmonary anastomosis. *J Thorac Cardiovasc Surg* 1996;111:1169-76.
9. Vitullo DA, DeLeon SY, Berry TE, et al. Clinical improvement after revision in Fontan patients. *Ann Thorac Surg* 1996;61:1797-804.
10. Mavroudis C, Backer CL, Deal BJ, Johnsrude CL. Fontan conversion to cavopulmonary connection and arrhythmia circuit cryoablation. *J Thorac Cardiovasc Surg* 1998;115:547-56.
11. McElhinney DB, Reddy VM, Moore P, Hanley FL. Revision of previous Fontan connections to extracardiac or intraatrial conduit cavopulmonary anastomosis. *Ann Thorac Surg* 1996;62:1276-83.
12. Abella RF, Marianeschi SM, De la Torre T, et al. The conversion of a modified Fontan procedure to a total extracardiac cavopulmonary conduit. The Medico-Surgical Cardiology Group [in Italian]. *G Ital Cardiol* 1998;28:645-52.
13. Lardo AC, Webber SA, Friehs I, del Nido PJ, Cape EG. Fluid dynamic comparison of intra-atrial and extracardiac total cavopulmonary connections. *J Thorac Cardiovasc Surg* 1999;117:697-704.
14. Amodeo A, Galletti L, Marianeschi S, et al. Extracardiac Fontan for complex cardiac anomalies: seven years' experience. *J Thorac Cardiovasc Surg* 1997;114:1020-31.
15. Petrossian E, Reddy VM, McElhinney DB, et al. Early results of the extracardiac conduit Fontan operation. *J Thorac Cardiovasc Surg* 1999;117:688-96.

Discussion

Dr John E. Mayer (*Boston, Mass*). The management of patients with a failing Fontan circulation may become an increasingly important problem for us over the next several years. We now have an ever-increasing cohort of patients who have survived a Fontan-type operation, and yet the natural

history of the patient with a Fontan circulation is still being written. I certainly congratulate you on bringing this combined experience from four institutions to us and bringing this topic onto the agenda for us to consider. I have a few points that I thought would be important to address.

One is the issue of postoperative arrhythmias. What role do you currently think the maze procedures or modified maze procedures or other antiarrhythmic types of operations would have in future management of similar kinds of patients?

My second question probably reflects the local custom in our institution regarding fenestration: Do you think there is any significant role for fenestration as part of a conversion from a so-called classic Fontan operation to a more recent version?

The third area in which I have a question, and I think it is always hard to determine this when you have patients who have multiple indications for an operation, is that of protein-losing enteropathy. We have certainly observed patients who had relatively favorable hemodynamics in terms of right atrial pressures and still had protein-losing enteropathy. We have wondered again recently about the role of lymphatic dysfunction in the origin of this kind of problem. Do you think this protein-losing enteropathy is simply attributable to the hemodynamics of the Fontan circulation or might other factors be contributing?

Dr Marcelletti. Thank you very much. I will answer questions two and three, and I will ask Constantine Mavroudis to give his contribution on the arrhythmia issue, since he has been the most active in this field.

We do not have any strong feeling against fenestration. If coming off cardiopulmonary bypass is somewhat difficult, fenestration will be considered. It is of interest that we did not find any need to fenestrate in this group of patients.

With regard to protein-losing enteropathy, we do not have much experimental data, but we have some clinical data. We have reported separately in *The Annals of Thoracic Surgery* the results of 3 patients who completely healed after conversion. We believe that protein-losing enteropathy has much to do with the energy loss and the turbulence in the right atrium, which slows the flow even when the mean right atrial and pulmonary artery pressures remain in the range of 15 to 18 mm Hg. Protein-losing enteropathy may be treated by conversion to an extracardiac Fontan, simply because the flows in the inferior vena cava, right atrium, and pulmonary artery improve, becoming more laminar.

Dr Constantine Mavroudis (*Chicago, Ill*). I would like to congratulate Dr Marcelletti for a remarkable synthesis of data from different institutions. Dr Mayer, your comments are very well taken. Our profession is following a large group of atriopulmonary Fontan patients who are getting older and who are now having significant atrial arrhythmias, among other conditions. Recent modifications of the Fontan operation incorporating lateral tunnel and extracardiac techniques may limit the incidence of atrial tachycardia, as shown in preliminary studies by Dr Laks and others. I suspect, however, that it is only a matter of time before troubling tachycardias affect these patients as well.

The potential for future atrial arrhythmias, anatomic obstructions, and other structural problems in these patients is high. Initial reports by Drs Laks, Kreutzer, McElhinney, and Vitullo extolled the virtues of the "redo atriopulmonary to total cavopulmonary artery conversion without arrhythmia" operation to treat the troubling anatomic and electrophysiologic problems that some of these patients face. Unfortunately, their results did not show consistent ablation of atrial arrhythmias. Our approach is to identify Fontan patients with troubling arrhythmias and/or anatomic abnormalities and consider an "extracardiac total cavopulmonary artery Fontan conversion with arrhythmia" operation, consisting of a modified right-sided maze procedure or a Cox maze III procedure in the event that the patient has atrial fibrillation. An atrial pacemaker is also inserted to maintain atrioventricular conduction. We have now operated on 22 patients who had Fontan conversion and arrhythmia surgery without short- or long-term mortality. Late postoperative arrhythmias occurred in two patients who are receiving long-term antiarrhythmic medications. All patients, except one who is on the transplant list for persistent left ventricular failure, are in NYHA class I or II. We believe that total cavopulmonary artery Fontan conversion in association with modified right-sided maze or Cox maze III procedures and pacemaker placement can be accomplished with low morbidity and mortality and results in functional class improvement and control of life-threatening arrhythmias.

Dr Antonio Amodeo (*Rome, Italy*). I have two questions.

You have been using, presumably, an aortic homograft for the extracardiac tunnel. We have done more than 120 extracardiac conduits, and we have used the aortic homograft as a lateral tunnel in three. One of these three homografts needed to be replaced. Another one is going to be replaced because of heavy calcification of the aortic wall causing a stenosis proximal to the pulmonary arteries. This kind of stenosis cannot be treated by balloon dilatation because of the heavy calcification of the homograft, whereas we currently use balloon dilatation for the polytetrafluoroethylene tube. Do you have any similar experiences when using aortic homografts? We no longer use aortic homografts to construct an extracardiac tunnel.

Second, you used a reconversion for one or two patients in NYHA class I. Do you think that all of the old aortopulmonary connections should be reconverted in an extracardiac tunnel? What are the early criteria to reconvert this old aortopulmonary connection?

Dr Marcelletti. With regard to the homograft, we are aware that some have calcified in the setting of an extracardiac Fontan conduit. The choice of the optimal conduit is still uncertain. In the opinion of most surgeons with experience in such operations, the polytetrafluoroethylene tube is a reliable conduit.

The indication to convert to extracardiac Fontan is based mostly on the enlargement of the right atrium, the so-called giant right atrium. It might not be giant, but it might already be enlarging. Such enlargement will shortly be accompanied

by symptoms such as arrhythmias, exercise intolerance, and protein-losing enteropathy. I think the basic alarm for taking down the previous Fontan conduit is the size of the right atrium.

Dr Joseph J. Amato (*Chicago, Ill*). Perhaps I am missing a point. I noted that you only had one lateral tunnel failure. I do not think you told us the reason for that lateral tunnel failing.

Like many of us who do lateral tunnels, I am concerned that in the growing child, if an extracardiac conduit is used, there is no other alternative but to replace the conduit as the child grows. Why not convert these from giant atria to lateral tunnel formations rather than extracardiac conduits?

Dr Marcelletti. The problem related to the lateral tunnel was thrombosis of the tunnel, which required revision to an extracardiac connection.

With regard to conversion of the previous Fontan, most of these patients have reached almost adult age, so we believe that the conduit will not need to be replaced. If the inferior vena cava remains around 20 to 25 mm, it should be all right. So far, none of the more than 200 patients with an extracardiac Fontan connection has required conduit replacement.

Dr Hillel Laks (*Los Angeles, Calif*). I have two questions: First, what was the arterial oxygen saturation of these patients before and after the operation? Second, what type of anticoagulation are you recommending over the short and long term for the extracardiac conduit?

I have one comment with regard to the fenestration. As opposed to the usual first Fontan procedure, during which the patients are cyanotic, these patients have saturations that are usually 92% to 94%. I would be interested to hear what your patients' saturations were.

We have found that in the revision we have made connec-

tions between the extracardiac conduit and the right atrium, either a direct window or a short polytetrafluoroethylene graft, and have found that these patients tolerate desaturation far less well than the first-time Fontan procedures. Some of our patients have become acidotic with saturations getting down into the 80% range. It is very useful in these patients to have an adjustable snare around the connection so that if the saturation is too low, the size of that communication can be readjusted.

Dr Marcelletti. I agree with you that fenestration might induce the problem of desaturation. We should not have any prejudice against fenestration in a patient who is having difficulty in being weaned from bypass, but I also believe that we should aim at full oxygen saturation after surgery.

Dr Laks. And the anticoagulation regimen?

Dr Marcelletti. We have been using warfarin sodium (Coumadin) for the first year after conversion. In the beginning of our experience, we monitored our patients with magnetic resonance imaging to see whether there was significant neointima deposition inside the conduit. We have observed only a small reduction in the inside diameter of the conduit. After 1 year, we convert to aspirin therapy.

Dr Laks. We recently did a transplant on a 17-year-old patient who had had an extracardiac Fontan revision 2 years earlier and was maintained on warfarin. There were patches of pseudointima built up and calcification within the polytetrafluoroethylene tube. I was a bit worried about the long-term outcomes and the question of whether warfarin combined with antiplatelet agents should not be used for a longer term, but there were patches of calcification within this tube.

Dr Marcelletti. I think that calcification and thrombus deposition inside the conduit after surgery has much more to do with the low output state than with the mode of anticoagulation.